We describe the case of a 48-year-old patient presenting with abdominal pain with a history of cerebral ischemia due to a patent foramen ovale with heterozygous factor V mutation. Initial work-up demonstrate a significant thrombosis of the portal venous system combined with signs of portal hypertension (ascites, oesophageal varices). Ultrasound reveals no signs of cirrhosis of the liver. Finally a JAK2 mutation can be detected. Prevention of oesophageal varices is refused. Finally a massive haemorrhage occured.